

# Myelodysplasia

## Problems of Long-Term Survival and Social Function

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*Problems of ninety-eight patients with myelodysplasia, ages 13 to 72, were reviewed. They were grouped as follows: Those having thoracic and high lumbar level ( $L_2\uparrow$ ) lesions and confined to wheel chairs, those with intermediate paralysis ( $L_{3-5}$  nerve roots) as walking with aids and those with less paralysis ( $S_1\downarrow$ ) as fully ambulatory. Fifty-two percent of the  $L_2\uparrow$  and only 15 percent of the less severely paralyzed patients were retarded below an IQ level 70 ( $P<0.01$ ). Thirty-six patients (62 percent) were fully and 26 partially, but appropriately, self-sufficient. Thirty-six patients were found in some form of dependent care. Two of the 71 more paralyzed patients ( $L_{3-5}$  and  $L_2\uparrow$ ) and five of the 28  $S_1\downarrow$  patients were "naturally continent" but reported stress incontinence of urine. Thirteen of 23 female and five of 28 male patients between ages 16 and 72 years reported sexual activity and accounted for 17 normal offspring. All 23 retarded patients were in some form of custodial care. Dependency among the normal intellect patients could be attributed to neglect of physically deforming complications and emotional disorders, primarily low self-esteem centering around social and sexual identity problems associated with excrement soiling.*

THE LITERATURE CONTAINS little information about teenage or adult patients who were born with myelodysplasia. In 1952 Fisher et al reported<sup>1</sup> survival of 471 myelodysplasia patients,

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Submitted May 7, 1974.

Supported in part by Clinical Research Center Grant No. RR-37, Research in Child Development and Mental Retardation Grant No. HDO2274 and aided by grants from The National Foundation-March of Dimes.

Presented in part before the Canadian Urological Society, Vancouver, British Columbia, June 13, 1973, and the 4th International Conference on Birth Defects, Vienna, September 4, 1973.

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17 percent of whom were between the ages of 20 and 53 years. They did not mention functional status. Four groups of investigators<sup>2-5</sup> have described the functional status of 83 adults and teenagers with congenital paraplegia. Most were under-achieving in school, in employment and in performance of self-help skills when compared with patients who had similar degrees of acquired paralysis. Genetic studies report four myelodysplastic children have been born among 114 children of 226 living myelodysplastic adults.<sup>6,7</sup> These latter two studies imply a relatively effective social adaptation but contain no descriptive details.

# MYELOYDYSPLASIA

TABLE 1.—*Intellectual Performance*

Functional Spinal Neuromuscular Level	DEGREE OF MENTAL RETARDATION					
	Profound to Moderate	Mild	Borderline	Normal	Bright Normal	Superior and Very Superior
	RANGE IQ SCORES					
	<51* <54†	52-68* 55-69†	69-83* 70-84†	84-110* 85-109†	111-120* 110-119†	121* and > 120† and >
L <sub>2</sub> ↑ (N=27)	9(33%)	5(19%)	4(15%)	8(30%)	0	1(3%)
L <sub>3-5</sub> (N=43)	4(10%)	1(2%)	8(20%)	22(50%)	5(12%)	3(6%)
S <sub>1</sub> ↓ (N=28)	2(7%)	2(7%)	6(21%)	13(47%)	3(11%)	2(7%)

( ) = % of patients by level of lesion.  
 \*SB = Stanford Binet.  
 †WISC = Weschler Intelligence Scale for Children.

TABLE 2.—*Summary of Social and Educational/Occupational Function for Myelodysplasia Patients With IQ Greater Than 70*

Functional Status	Level of Lesion*	Age in Years		
		13-18†	19-26‡	Older than 27 Age in ( )
Fully employed or housewife with complete self-help skills	L <sub>2</sub> ↑	0	3	1 (51)
	L <sub>3-5</sub>	0	2	2 (30 & 73)
	S <sub>1</sub> ↓	0	3	2 (34 & 34)
Normal grade in school or in higher education & complete self-help skills	L <sub>2</sub> ↑	0	0	0
	L <sub>3-5</sub>	10	3	0
	S <sub>1</sub> ↓	8	2	0
Living at home or in custodial care with partial self-help skills and in special education or job training	L <sub>2</sub> ↑	6	1	0
	L <sub>3-5</sub>	11	2	0
	S <sub>1</sub> ↓	4	2	0
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Custodial care or living at home with no program	L <sub>2</sub> ↑	0	2	0
	L <sub>3-5</sub>	1	5	2 (57 & 34)
	S <sub>1</sub> ↓	0	2	0

\*See text for definition.  
 †Age range eligible for grade or high school.  
 ‡Age range eligible for college and higher education.

Most observers reporting upon older patients describe a group of patients referred to their institutions for rehabilitation—that is, selected. We have studied nearly all living myelodysplastic patients in our area (we were not able to trace three patients), regardless of the degree of handicap. The purpose of this report is to describe the intellectual capabilities, neuromuscular function, educational and economic status, and social adaptation of 98 myelodysplastic patients between the ages of 13 and 73 years.

## Method

Patients born with myelodysplasia (for example, spina bifida aperta, myelomeningocele, meningomyelocele) were brought into this study through record reviews and health department

birth records. Most have been followed for up to 17 years in a multiple disciplinary clinic. All clinical data have been recorded on standardized record forms and analyzed for survival using the conversational computer statistical service.<sup>8</sup> We have derived information concerning the presence and treatment of hydrocephalus, intellectual performance and various types of self-help skills from these computer records (28 items). Current social and economic or educational status were added. Intellectual assessments were obtained by a battery of gauges including the Weschler Intelligence Scales for Children (and Adults), Wide Range Achievement and Stanford-Binet tests.

Grading of muscle strength was recorded according to the Sharrard classification.<sup>9</sup> Grouping of our patients was made by functional motor activity as follows:

- Lesions at or above the second lumbar vertebra (L<sub>2</sub>↑)—All thoracic and lumbar root levels 1 and 2—intercostal, abdominal and hip flexion and adduction muscles function—essentially all wheelchair patients.

- Lesions between the third and fifth lumbar vertebrae (L<sub>3-5</sub>)—Additional muscle strength in quadriceps and hamstrings giving stronger hip flexion and adduction with knee flexion (weak) and extension; some patients have anterior tibialis with weak foot dorsiflexion and weak hip abductors—most walk effectively with aids, surgical operation and extensive bracing.

- Lesions at or below the first sacral vertebra (S<sub>1</sub>↓)—Additional muscle strength about the ankle with variable degrees of hip abduction and extension—essentially all walk with minimal aids.

Confirmation of sexual activity for women was based on history, with the majority confirmed by pregnancy. Confirmation of male activity was accepted only with corroborating evidence.

Educational or occupational and social func-

# MYELOYDYSPLASIA

TABLE 3.—Reasons For Failure to Achieve Expected Goals—  
Custodial Care Without Program (12 Patients)

Level of Lesion	No. Patients	Age In Years	Sex	Untreated Complications	IQ	Major Cause for Failure	Other Comments
L <sub>2</sub> ↑ . . . . .	2	20*	♀	Yes	105	Scoliosis, decubiti, urine soilage	Family rejection
		20	♀	Yes	75	Family rejection and personal psychological problems	Obesity, poor motivation
L <sub>3-5</sub> . . . . .	8	18†	♂	No	90	Family psychopathology, disturbed child with poor motivation	Urine soilage, school dropout
		19†	♀	Yes	80	Early educational deprivation, family rejection, obese	Married to mental retardate, wheelchair, IQ=61 at age 14 yrs; after special educ., employed as telephone operator briefly
		24†	♂	Yes	112	Psychosexual disturbance, condom drainage with poor hygiene	Penile slough, lack motivation
		24	♀	No	70	Psychosexual pathology, sexually permissive	.....
		24†	♀	No	95	Psychosexual pathology, family psychopathology	Unwed mother
		24	♀	No	70	Psychosexual pathology	Sexually active, school dropout
		34†	♂	Yes	122	Psychosexual pathology, lack motivation	Dropped out of work despite graduate degree; sexual fantasy life
		57†	♂	No	127	Coronary artery disease, celibate, over-concerned about hygiene	Lost position, now unable to find employment despite graduate degree
S <sub>1</sub> ↓ . . . . .	2	21†	♂	Yes	70	Family rejection	Married, both partners with venereal disease, patient's child put in custody
		22	♂	Yes	115	Dying of renal failure, lacks motivation, psychosexual pathology	Urine soilage

\*Only patient totally bedridden. All others are capable of feeding, dressing, personal hygiene and self mobility.  
†Living at home on welfare subsidy.

tion were defined as independent or custodial living, complete or appropriate partial (for example, teenagers too young to drive), or no self-help skills and normal grade placement in school or employment, or special education, sheltered workshops or special job training.

## Observations

Complete information, examinations, and tests were obtained for 98 patients. Three additional patients could not be contacted for this study.

## Intelligence

Intellectual performance has been recorded in Table 1. Fifty-two percent of patients with L<sub>2</sub>↑

level lesions were classified in the profound to moderate or mild retardation group as compared with only 15 percent of the lower level lesion patients ( $X^2 = 12.8$ ,  $df = 1$ ,  $P < 0.01$ ) and 6 percent of the general population.

## Occupational or Educational and Social Status

A summary of the occupational or educational and social status of the patients with borderline, normal, or better than normal intellect has been recorded in Table 2. Among 36 patients who are completely self-sufficient and in either normal academic level or fully employed, there are ten with severe organic or social adjustment complications (above solid line in Table 2). One father

# MYELOYDYSPLASIA

TABLE 4.—Reasons for Failure to Meet Expected Goals—Living at Home or in Custodial Care in Special Education or Job Training (11 Patients)

Level of Lesion	No. Patients	Age	Sex	Capable of Self-Help Skills	Untreated Complications	IQ	Major Cause for Under-performance	Other Comments
L <sub>2</sub> ↑	4	13	♀	Partly	Yes	97	Family psychopathology, renal failure	Scoliosis, obesity, lack of motivation
		14	♀	Partly	Yes	90	Family psychopathology	Dislocated hips, recurrent fractures of legs, poor urine hygiene
		16	♀	Partly	No	75	Lack of motivation; three CNS infections	Scoliosis
		19	♀	Partly	Yes	75	Huge head, renal hypertension	Recurrent decubiti, lack of motivation, denied early Rx
L <sub>3-5</sub>	5	13	♀	Yes	No	72	Seizures, poor family situation	Foster care, low intellect, divorced parents
		16	♀	Partly	No	70	Family rejection	Urine soilage, placed in an institution
		17	♀	No	No	77	Spastic quadriplegia	Excellent care—medical, school and home; severely handicapped following shunt obstruction and CNS infection
		19	♂	Yes	Yes	70	Neurologic sequelae of untreated hydrocephalus	Severe decubiti and osteomyelitis, secondary to urine soilage; family rejection
		21	♂	Yes	No	77	Family rejection	Urine soilage
S <sub>1</sub> ↓	2	13	♂	Partly	No	70		
		14	♂	Partly	No	70		Urine soilage

with three children is dying of renal failure.\* Severe decubitus ulcers and osteomyelitis developed in an unmarried young man causing reduction in his locomotor status from ambulation with Lofstrand crutches to a wheelchair. Both of those patients had sacral level lesions. Three unmarried young men have serious emotional problems, in part relating to urine incontinence and difficulty with sexual adjustment. Two young women have been disappointed by broken engagements and one has been left an unwed mother. Two other young women, though interested in men, are ineffective in relating to them. The other 26 appear to be well adjusted and are either making good progress toward or have achieved social stability and self-sufficiency.

Twelve patients (below the dashed line in Table 2) have failed to perform in an expected social, occupational or educational manner. Reasons for failure, usually multiple, are recorded in Table 3.

\*Patient died eight months after study was completed.

Twenty-six patients (ages 13 through 26) were intermediate between the above two groups because of (1) young age, or (2) they are still in an educational or training status and (3) because of a low intelligence quotient. Eleven of these 26 are not expected to succeed, for reasons stated in Table 4. Considering their handicap, the others were progressing well in school or special education and medical rehabilitation programs. If their progress continues, they can be expected to become fully self-sufficient.

## Urine Control

Only one female patient among the 27 patients with L<sub>2</sub>↑ and one among the 43 with L<sub>3-5</sub> level lesions have developed socially acceptable natural continence of urine. Two female and three male patients among 28 with S<sub>1</sub>↓ lesions (17 percent) have some degree of urinary continence. However, frequent stress incontinence causes social embarrassment for most of these seven pa-

# MYELOYDYSPLASIA

TABLE 5.—Method of Urine Collection, Teenage and Adults, 1973

	<i>Socially Satisfactory</i>	<i>Socially Unsatisfactory</i>	<i>Biologic Complications</i>
Early Ileal Diversion	28(93)†	0	2
Late Ileal Diversion	4(28)	0	10
MR and Diaper	14(82)	0	3
Natural Drain	6(86)	0	1
Napkin/Diaper	2(25)	5	1
Sheath	5(63)	2	1*
Clamp	1(50)	1	0
Catheter	3(60)	1	1*
Ureterostomy	0	0	1
Suprapubic	0	0	2*

\*One each now with satisfactory loop.

†Figures in ( ) are percentages.

tients. Methods of urine collection for the other 91 known patients have been recorded in Table 5. Satisfactory urine control was best in patients who had ileal diversion early. Recurrent urinary tract infections, stone formation and the persistence or progression of hydronephrosis occurred following diversion only where the diversion was not performed until after the onset of hydronephrosis and ureterectasis.

## Sexual Activity

Sexual activity of 51 patients between the ages of 16 (youngest patient involved) and 72 years is recorded in Table 6. Eighteen of these 51 patients were judged to be participating in sexual activity. Several with colorful but unconfirmed stories were not included. Female patients had premarital sexual relations more frequently than males ( $P < .01$ ). Both male and female patients described varying degrees of gratification and pleasure from sexual intercourse. Only one woman described occasional painful or unpleasant sensations with coitus. Although only one of the five involved men experienced erection, manual penile manipulation at the introitus provided adequate satisfaction. One sired three children. All of the 17 offspring from these myelodysplastic parents have been normal. Each of the two women with ileal diversion who have borne a child requested tubal ligation because of urinary tract infections, urinary obstruction, and ileal loop stoma difficulties in the third trimester.

## Social Attainment of Retarded Patients

Social function and educational status for 23 retarded myelodysplastic patients ages 13-22 years were limited. None of these 23 patients shows evidence of being able to achieve an independent social life. Eighteen are profoundly to

TABLE 6.—Sexual Activity in 51 Myelodysplastic Patients, Age 16-72

	<i>Male</i>	<i>Female</i>	<i>Totals</i>
<i>No Sexual Activity:</i>			
Too handicapped	23	10	33
Virginal	14	4	18
<i>Sexual Activity:</i>			
Premarital	9	6	15
Marital	5	13	18
Both	1	8	9
TOTALS	3	3	6
	1	2	3
	28	23	51

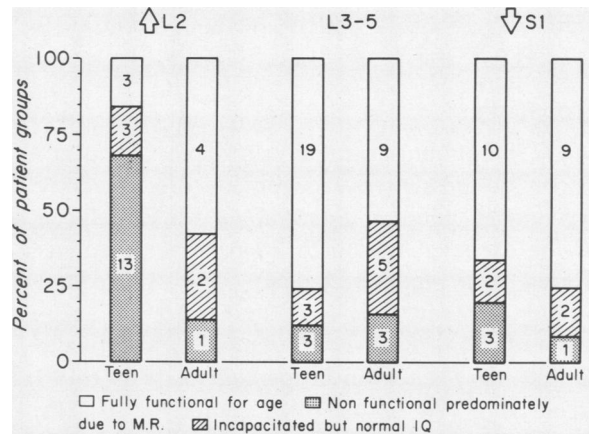


Chart 1.—Attainment of teenage patients with myelodysplasia compared with that of adult patients.

severely retarded (IQ of 10 to 50). Ten of these had L<sub>2</sub>↑ level lesions. All 18 patients have a combination of physical deformity and low intellect that render them bed-care patients. The other five patients, less retarded, were in custodial care but had partial self-care abilities and were engaged either in special education or job training (IQ of 51 to 66). Only one of the latter group is not in the L<sub>2</sub>↑ level lesion group. Total custodial care costs \$6,500 a year, whereas partial to complete self-care reduces the cost to an average of only \$1,500 a year.

## Comparison of Teenage and Adult Attainment

Chart 1 compares the attainment for teenage patients (13 to 18 years of age) with that of adults (19 years and older). Approximately the same proportion of teenagers as adults with S<sub>1</sub>↓ lesions are fully functional for age, are incapacitated physically but with normal IQ, or are in custodial care. Eighty-four percent of the high level lesion (L<sub>2</sub>↑) teenagers are in custodial care due to physical disabilities, mental retardation, or both. This proportion of non-functional teenagers is twice that of similarly paralyzed older patients

(.02 < P < .05). That more teenagers than adults in the L<sub>3-5</sub> paralysis group are fully functional is of questionable significance (.05 < P < 0.1). In the six-year teenage lifespan there are many more survivors (59) than the 41 adults representing the older seven decades of life (X<sup>2</sup> = 44, df = 1, P < 0.001). Four (10 percent) adults (1 with L<sub>2</sub>↑ and 3 with L<sub>3-5</sub> level paralysis), and 15 (26 percent) teenagers (9 with L<sub>2</sub>↑, 5 with L<sub>3-5</sub> and 1 with sacral level lesions), have shunt treated hydrocephalus (X<sup>2</sup> = 3.2, .05 < P < 0.1). Eight of the 19 with shunts are retarded (42 percent) compared with 15 of 79 patients without shunts (19 percent) (X<sup>2</sup> = 3.4, df = 1, .05 < P < 0.1).

## Discussion

Few infants born with myelodysplasia and active or progressive hydrocephalus are currently surviving as adults.<sup>15</sup> Survival to adulthood can now be expected for 60 to 70 percent of infants referred to a regional center.<sup>14,15,29,30</sup> There are, however, too few long-term studies for an accurate prediction. Survival of treated hydrocephalic children has risen from 60 percent in 1963 to 90 percent in 1974 in our own center.<sup>31,32</sup> Earlier referral, more appropriate selection, more effective spinal fluid shunt systems, improved understanding of shunt complications and treatments, as well as availability of broader spectrum antibiotics may account for this better survival. Hence, of the one myelodysplastic baby in fifteen hundred births in the United States, probably 50 percent will survive to adulthood through use of currently accepted methods of treatment.

Even without early intervention, a number of myelodysplastic patients have survived to adulthood with varying degrees of lower extremity paralysis. Many have no evidence of hydrocephalus although some have enlarged heads or a (poorly documented) history of rapid head growth in infancy. Our 41 adults and most of our older teenagers have managed to survive with arrest of hydrocephalus in a way comparable to that of other patients mentioned in the literature.<sup>1-5</sup> High level motor lesions observed in these older patients may have been originally low level motor lesions that became locally infected, traumatized, or scarred. The more common occurrence of mental retardation among those with high level lesions is probably associated with cephalad neural tube dysplasia that is coincident with failure of the caudal neuropore to close.<sup>10,11</sup> Lower level lesions may occur more frequently from coales-

cence of distal neural elements<sup>12</sup> and, therefore, may be less frequently associated with cephalad dysplasia and mental retardation.

Irrespective of types of reconstructive and educational care provided in later life, primary brain malformation may explain the increased proportion of mental retardates in our young teenage population and in the studies of other young patients.<sup>4,13-16</sup> The data in this study only suggest that the increase in survival of myelodysplastic, teenage patients is related to the survival of a greater proportion of mentally retarded, shunt-dependent patients.

Untreated, neuromuscular imbalance across joints can cause contractures and scoliosis that impede or prevent self-mobility. Asymmetric weight-bearing secondary to contractures as well as neurogenic anesthesia can cause severe perineal decubitus ulcers. The incidence of ulcers increases considerably with excretory soiling.<sup>17,18</sup> Untreated decubitus ulcers, by extending infection to soft tissue, bone and joint have reduced some of our patients from walkers to wheelchair or bed existence.

In addition, myelodysplastic patients suffer from emotional consequences of incontinence.<sup>3,5,17,18</sup> Emotional disturbances stem from low self-image, excreta soilage, and odor ("outhouse syndrome"), poor motivation, lack of opportunity for heterosexual intimacy and poor sexual identity. Congenitally paraplegic patients can enjoy sexual activity and reproduce just as do those whose paralysis is caused by trauma.<sup>5-7</sup> Some exaggerate their sexual prowess even more than do their "normal" peers of the current culture. Regardless of the degree of fact or fancy, management of urine collection should take into consideration the need for sexual adaptation. Urine collection devices such as the penile clamp, sheath collector, Foley catheter and suprapubic cystostomy tube have led to severe physical complications secondary to adolescent rebellious misuse of them, as well as to deep-seated emotional scars.

Several female patients were inappropriately advised about their fertility, and unexpected pregnancy followed. Appropriate reproductive counseling should include five separate concepts:

- Male and female paraplegic patients describe gratification as a result of sexual play.<sup>5</sup>

- Female paraplegic patients can conceive.<sup>6,7</sup> In the absence of complicating infections that could produce secondary sterility and despite erectile impotence, adult male paraplegic patients are fertile, if they ejaculate.<sup>6,7</sup> It is possible for a male

patient born with a flail penis to have insertion of an artificial, prosthetic corpora erecta<sup>19</sup> and an internal urinary bladder sphincter<sup>28</sup> to prevent retrograde ejaculation.

- Approximately 3 percent of children born of a myelodysplastic parent are born with a neural tube defect.<sup>6,7</sup>
- Amniocentesis can identify affected fetuses.<sup>20</sup>
- A paralyzed patient with an ileal diversion, in our experience, has a difficult and hazardous third trimester of pregnancy.

Under-achievement is more common in the celibate male suffering from low self-esteem. This lower self-image is derived in good measure from physical impotence compounded by antisexual urine collection devices. Preoccupation with sex and an apparent compulsion to bear children to prove femininity can interfere with social adjustment of female patients.

The effect of mental retardation on the performance of older patients, and insidious urinary tract deterioration leading to irreparable hydronephrosis suggest the need for two types of preventive care early in the life of myelodysplastic patients. To provide for the highest possible quality and length of life, the patient must be protected against the sequelae of hydrocephalus and hydronephrosis. Modern advances in cerebrospinal fluid management<sup>21-23</sup> and control of shunt-related infections<sup>24</sup> provide the means for protecting the central nervous system against pressure effects and damage from infection. Frequent urinary tract evaluation and use of ileal diversion before the onset of hydronephrosis and hydroureter have preserved renal health of patients in this as well as in other series.<sup>25-27</sup> It may be hoped that urinary bladder outlet relaxing procedures combined with the insertion of a silastic, internal, extra-vesical artificial urine sphincter will provide even more aesthetic care for the neurogenic bladder in the future.<sup>28</sup>

Treatment of myelodysplastic patients should be directed at enhancing locomotion, aesthetic urine and stool control and self-care both for those with normal intellect and the mentally retarded. An infant with normal or potentially normal intellect should have life-sustaining treatment of potentially fatal central nervous system and urinary tract problems. A distinction should be made between the life-sustaining and life-improving merits of treatment. Procedures and treatments that reduce the patient's functional handicaps are ultimately both economically and morally sound.

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